Unnecessary laparotomy due to radiological misinterpretation

Ramos y Muñoz, G ; Moehrlen, U ; Bode, P K ; Arlettaz Mieth, R

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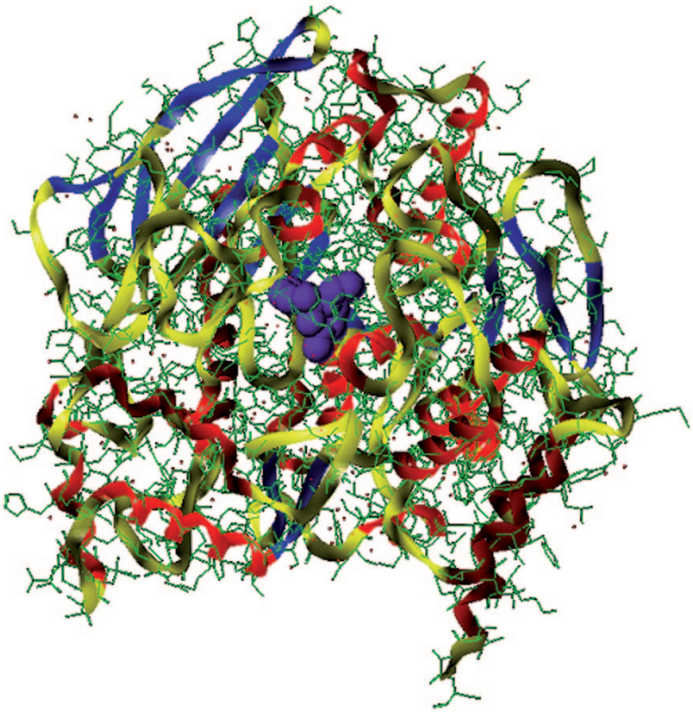

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Ramos y Muñoz G, Moehrlen U, Bode P, Arlettaz Mieth R, Clinic of Neonatology (RMG, AMR), University Hospital Zurich, Division of Surgery (MU), University Children's Hospital Zurich, Division of Pathology (BP), University Hospital Zurich

A male infant was born in a level I hospital at 40 2/7 weeks of gestation by caesarean section due to pathological cardiotocogram. The mother was a 36-year-old G4/P4. The family history was unremarkable. The three older children (29, 18 and 13 years of age) were reportedly healthy. The pregnancy had been uneventful, except for non-insulin-dependent gestational diabetes. The newborn had an umbilical arterial cord-pH of 7.24, base excess of -4 mmol/L and adapted well with an Apgar score of 9, 9 and 10 at 1, 5, and 10 minutes respectively.

The initial postnatal course was uneventful with passage of meconium and normal breast feeding. However, at the age of 24 hours, passage of hard stool was observed and the baby developed abdominal distension and had one episode of bilious vomiting. He was transferred to the NICU at the University Hospital in Zurich for further investigations.

On admission at the age of 36 hours, the baby appeared well and was afebrile. Physical examination revealed a tensely distended abdomen with reduced intestinal peristalsis. Livid-reddish periumbilical discoloration and a putrid smell from the umbilicus were observed. The baby had mild tachypnea without further signs of respiratory distress and without episodes of apneas. Blood pressure was normal. Blood gas showed normal values, lactate was slightly elevated at 3.5 mmol/l, C-reactive protein was 8 mg/L and leukocyte count was normal.

At that point, the leading diagnoses were Hirschsprung's disease or meconium ileus. Shortly thereafter, however, a conventional ap abdominal radiograph was performed and revealed dilated bowel loops, the presence of stool, and intestinal pneumatosis indicative of necrotizing enterocolitis (NEC) (Fig. 1). The X-ray was not seen by a radiologist, but it was interpreted by both surgeons and neonatologists.

After consulting with the pediatric surgeons, it was decided to treat this presumed case of NEC conservatively. Oral feedings were discontinued, a nasogastric tube was inserted and parenteral nutrition was started. After obtaining blood cultures, antibiotic therapy with amoxicillin/clavulanic acid and gentamycin was started. The clinical course over the next few hours was stable. There were no stools, no rectal bleeding, and only one episode of bilious vomiting. Six hours later, follow-up abdominal x-rays were obtained (Fig. 2, 3). On these images, the diagnosis of NEC became less likely as the air bubbles appeared to be mixed with stool.

The baby became more uncomfortable with increasing abdominal distension and was transferred to the University Children's Hospital of Zurich for an exploratory laparotomy. Intraoperatively, there were markedly distended loops of bowel but no evidence of NEC. Hirschsprung's disease was now felt to be the likely diagnosis. No further surgical interventions were performed but the stool was intraoperatively mobilized by enemas. Later, a

series of contrast studies of the small and large bowel showed a decreased recto-sigmoid diameter suggestive of Hirschsprung's disease (Fig. 4).

Following intra- and postoperative enemas, large amounts of stool were passed, followed by rapid clinical improvement. Enteral feeding with breast milk was started and well tolerated. Normal defecation was achieved by repeated dilatation of the anal canal with Hegar dilators. One week later, aganglionosis was demonstrated on a colorectal mucosal biopsy confirming the diagnosis of Hirschsprung's disease (Fig. 5). The baby was discharged home 2 weeks later. The final surgical resection of the abnormal recto-sigmoid section via pull-through procedure was successfully performed at the age of 6 weeks.

Fig. 1

Abdominal X-ray at the age of 37 hours: possible intramural air in some bowel loops (asterisk).

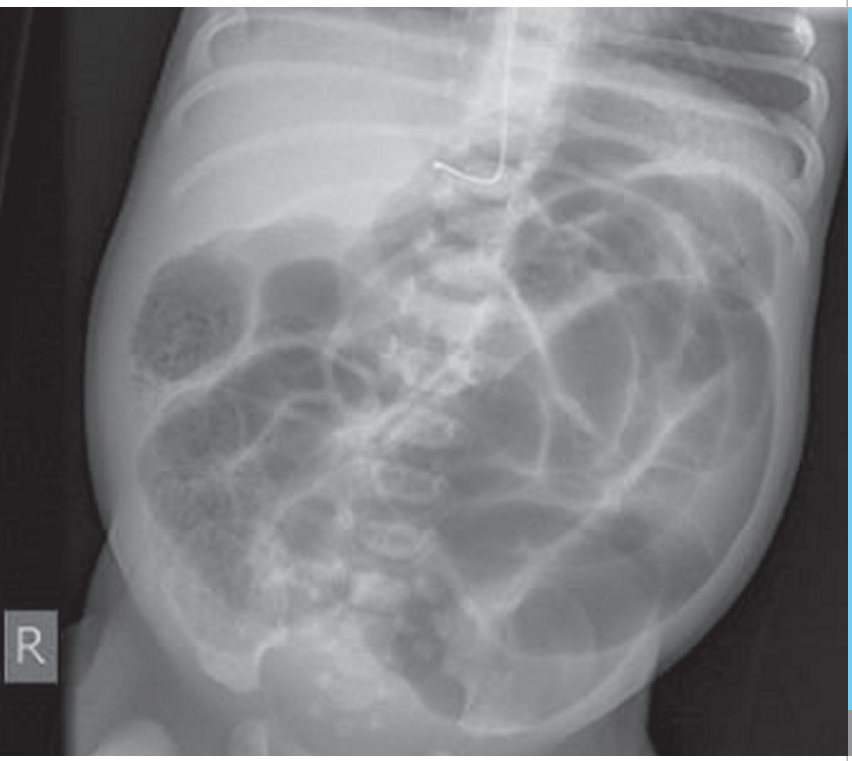


Fig. 2

Abdominal X-ray at the age of 43 hours: dilated loops of bowel and air bubbles which are not restricted to the bowel wall.

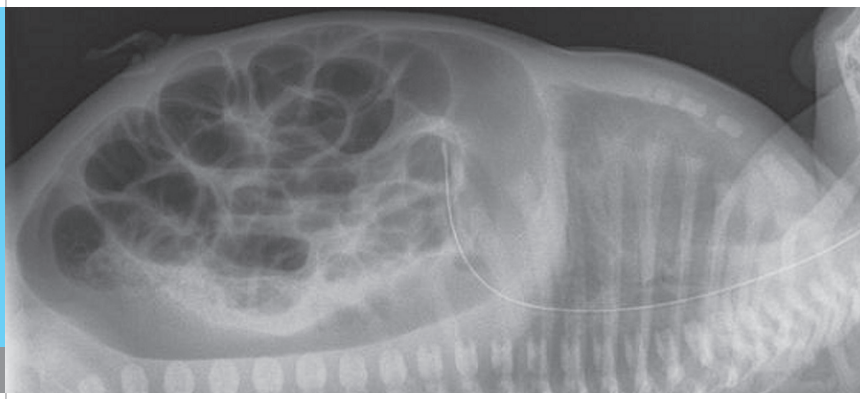


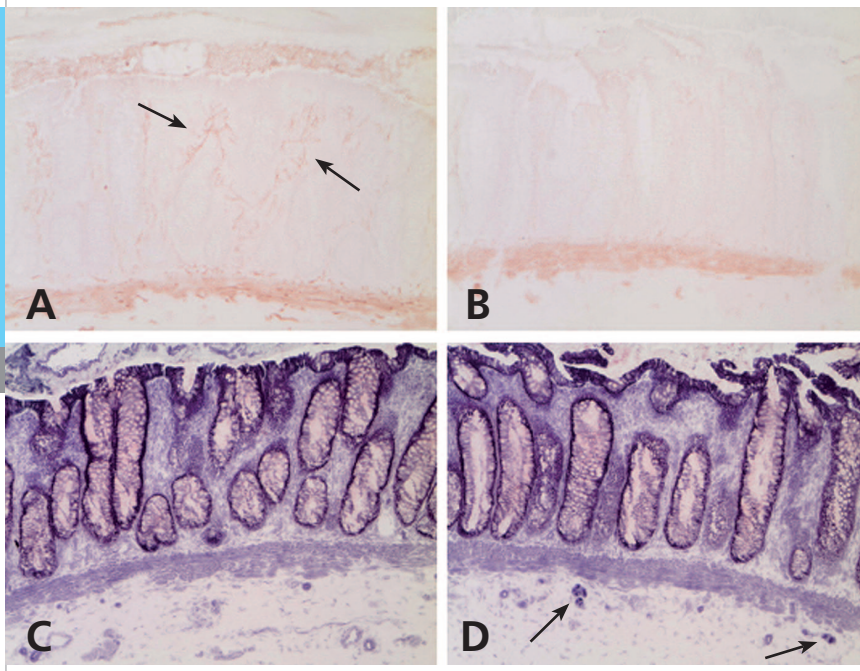
Fig. 3

Lateral abdominal X-ray at the age of 43 hours without evidence of perforation.

Fig. 4

Contrast study of the colon at the age of 9 days: narrowing in the region of the rectosigmoid.

Fig. 5



Histopathology of Hirschsprung's disease: A) Affected segment: multiple nerve fibers in the lamina propria with increased acetylcholinesterase activity (arrows); B) Normal colon without increased acetylcholinesterase activity (histochemical reaction for acetylcholinesterase on frozen sections); C) Affected segment: Absence of submucosal ganglion cells; D) Normal colon with normal distribution of ganglion cells in the submucosa (arrows) (histochemical reaction for nitric oxide synthase on frozen sections).

Hirschsprung's disease is caused by the absence of intramural ganglion cells in the lower digestive tract leading to a developmental disorder of the enteric nervous system. It is characterized by the interaction of multiple genes and manifests with low, sex-dependent penetrance and variability in the configuration of the aganglionic segment (1). In Hirschsprung's disease, the neural crest stem cells, which migrate along the intestine to establish the myenteric and submucous plexus, abort their migration during the embryonic development. This leads to a dysfunction of the organization of parasympathetic fibers, which grow into the distal colon. Gut overactivity with continuous contraction of the bowel due to a sustained release of acetylcholine is the result (2).

The incidence of Hirschsprung's disease is reported to be about 1:5'000 live births in both hemispheres (3). It commonly presents in the newborn period sometimes with life-threatening bowel obstruction requiring surgery.

The most prevalent gastrointestinal emergency in newborns is due to NEC which affects mostly extremely preterm infants. Without doubt, symptoms of Hirschsprung-associated enterocolitis (HAEC) and neonatal enterocolitis can be similar (4). Newborn infants with Hirschsprung's disease presenting soon after birth are at greater risk of developing postoperative HAEC. HAEC is associated with significant perioperative morbidity and mortality. Several studies on

HAEC have been published, however, the pathogenesis of this condition remains unclear (5).

In the neonate, pneumatosis intestinalis is almost always associated with NEC (6). Intestinal pneumatosis can be difficult to recognize on abdominal x-ray, or can be confounded with less relevant findings, like the presence of small gas bubbles within the stool filled bowel lumen, as was the case in our patient. Additional investigation with abdominal ultrasound can help to detect intestinal perforation related to neonatal enterocolitis. It has been shown that abdominal ultrasound is superior to plain radiography in identifying portal venous gas and echogenic free fluid as early signs of intestinal perforation (7).

Our patient developed signs and symptoms of ileus at the age of 24 hours. Although Hirschsprung's disease was felt to be the likely diagnosis based on history and clinical findings, radiological misinterpretation of intestinal pneumatosis lead to a diagnosis of NEC and explorative laparotomy. Early repetitive enemas would probably have led to rapid clinical amelioration and avoidance of surgery.

See also **COTM 03/2005:** Intestinal perforation in Hirschsprung's disease.

ACKNOWLEDGEMENT

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Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch